Case Report

A rare case of multiple cardiac rhabdomyomas

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Rhabdomyoma is the commonest benign cardiac tumor of childhood, although it rarely will be found in early adulthood. They originate within the myocardium, typically in the ventricles, and may be multiple in up to 90% of cases. Up to 50% of these hamartomas are associated with index cases of tuberous sclerosis. The majority of patients are asymptomatic, and most of these rhabdomyomas will spontaneously regress. However, some may produce life-threatening cardiac failure due to left ventricular outflow tract obstruction or arrhythmias, and these will require surgical resection.

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Key words **: Rhabdomyoma, cardiac tumor.**

Rhabdomyoma is the commonest benign cardiac tumor of childhood originating within the ventricular myocardium and may be multiple in up to 90% of cases. Up to 50% of these hamartomas are associated with index cases of tuberous sclerosis. The majority of patients are asymptomatic and most of these will spontaneously regress. However, some may produce cardiac failure due to left ventricular outflow tract obstruction or arrhythmias and require surgical resection. We present a case of multiple rhabdomyomas in a 20 days old male child, 2D echo suggested multiple solid hyperechoic varying size masses in bilateral ventricular free wall and interventricular septum. One of them was seen obstructing right ventricular outflow tract. All these findings were confirmed on cardiac CT.

Case Report

A 20 days old male child presented with complains of dyspnoea, cyanosis and feeding difficulty. So he was investigated and 2D echo was done to rule out underlying cardiac abnormality. 2D echo suggested multiple solid hyperechoic varying size masses in bilateral ventricular free wall and interventricular septum. One of the lesions was seen obstructing right ventricular outflow tract.

Patient had no history of seizures or any skin lesions. Family history was insignificant. Ultrasound of cranium and abdomen was unremarkable.

CT cardiac was also performed to rule out any other associated cardiac abnormality. CT was performed after injecting non-ionic contrast through antecubital vein. These images were reviewed with MIP (maximum intensity projection), SSD (shaded surface display) and Volume Rendering. CT Findings were multiple varying size non-enhancing filling defects that appeared isodense to myocardium involving bilateral ventricular free wall and interventricular septum. One of the lesions was projecting in right ventricular outflow tract. Hypoplastic confluent pulmonary arteries were seen.

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Discussion

A cardiac rhabdomyoma is type of benign myocardial tumor. Cardiac rhabdomyomas are often multiple and can represent up to 90 % of cardiac tumors in the pediatric population[1]. The majority are diagnosed before the age of 1 year. The estimated incidence is at ~ 1 in 20,000 births[4]. They may arise anywhere in the myocardium but are commoner in the ventricles (may involve the left ventricle more)[5].

Clinical presentation :

The majority of cardiac rhabdomyomas are asymptomatic although there can be wide clinical spectrum. On occasion they may present with ventricular outflow tract obstruction or refractory arrhythmias. There is a well known association with tuberous sclerosis, with over 50% of all cardiac rhabdomyomas found in patients with later confirmed tuberous sclerosis[2,3]. The remainder of these tumors occurs sporadically or in association with congenital heart disease. Although most infants with tuberous sclerosis have cardiac rhabdomyomas, the prevalence of these lesions in this population decreases with increasing age, because of spontaneous tumor regression and better survival of patients without cardiac tumors[1]. Other complications include valvular compromise or disruption of intra-cardiac blood flow leading to congestive heart failure and hydrops.

Pathology :

It is a hamartomatous lesion consisting of cardiac muscle tissue (derived from embryonal myoblasts). They grossly appear as yellow-tan solid, circumscribed, unencapsulated lesions. Microscopically, a characteristic spider cell is seen which a large clear cell is with cytoplasmic strands composed of glycogen extending to the plasma membrane[8].

Radiographic features :

Ultrasound / Echocardiography **—** May be seen as one or more solid hyper echoic masses located in relation to the myocardium. Small lesions can mimic diffuse myocardial thickening (Fig 1).

CT — Seen as filling defects iso to hypodense compared to myocardium involving ventricular wall or interventricular septum (Fig 2).

MRI — T1 : relatively well defined masses iso intense to adjacent myocardium

T2 : relatively well defined masses hyper intense to adjacent myocardium

Computed Tomography (CT) and Magnetic Resonance (MR) can provide very useful additional information for a better characterization of the tumor[6,7]. CT does not only facilitate the study of the heart but it also allows for the study of the adjacent mediastinum and extra cardiac anomalies. However CT does not provide real time images like the echocardiography and it is not useful for the analysis of valvular mobility. MR is better than CT for the characterization of soft tissues and is very useful for the evaluation of intramural masses. Rhabdomyomas detected with US may be missed with CT or MR imaging and vice versa. Thus, these procedures may be complementary. In general, US better demonstrates small (<0.5 cm) or entirely intramural lesions.

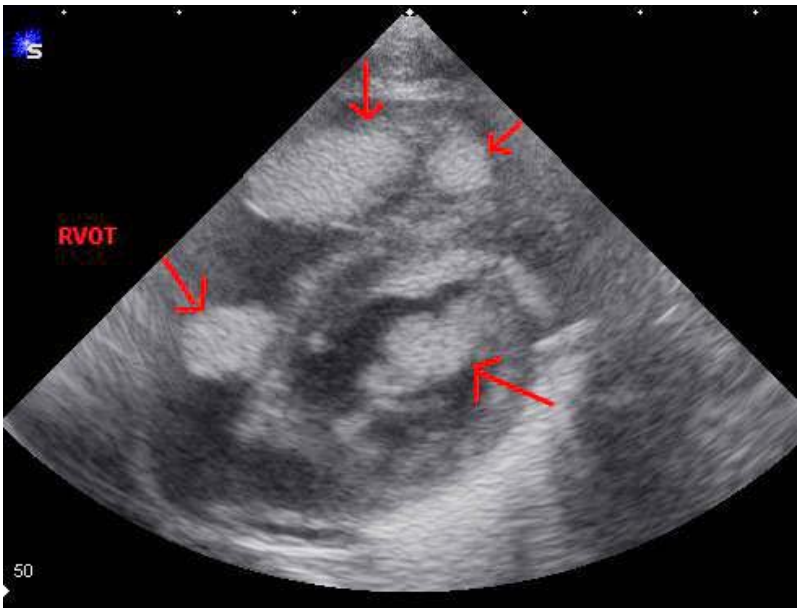

Fig 1 — 2D echo image showing multiple solid hyperechoic cardiac rhabdomyomas involving the ventricular septum and ventricular wall (arrows). One of the lesions is projecting in right ventricular outflow tract

Treatment and prognosis :

In most cases no treatment is required and these lesions regress spontaneously. Patients with ventricular outflow tract obstruction or refractory arrhythmias respond well to surgical excision. The overall prognosis is dependent on the number, size and location of the lesions as well as the presence or absence of associated anomalies.

Our patient was put on regular monthly follow-up to monitor any complications.

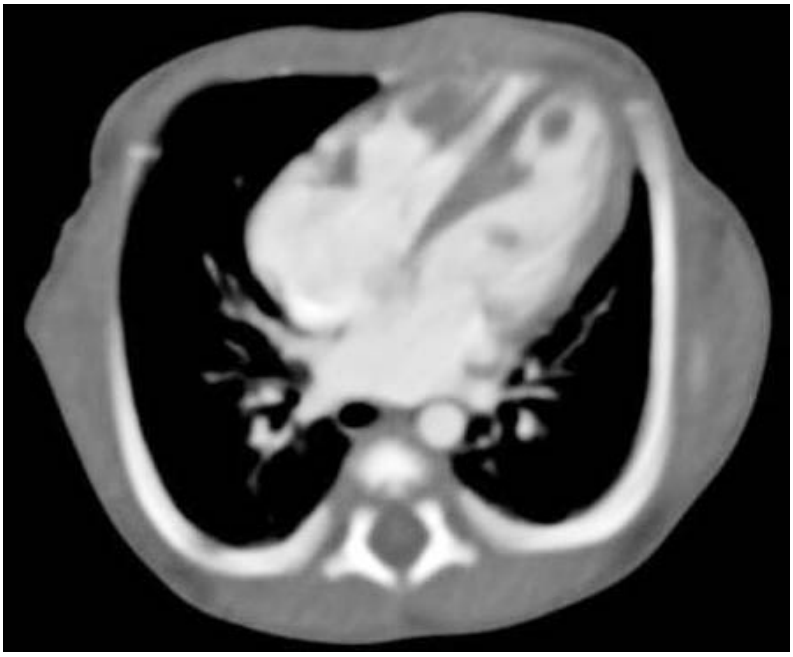

Fig 2 — CT image showing multiple non-enhancing filling defects involving bilateral ventricular free wall and interventricular septum

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